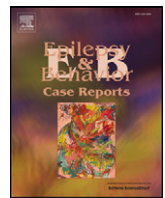


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Case Report

Combined variants in reading epilepsy; coexisting anterior and posterior variants camouflaged as heat cramps where the patient finds his own diagnosis searching the internet

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ABSTRACT

Reading epilepsy is a form of reflex-induced seizures. Two entities are postulated as part of a clinical spectrum; one anterior variant with jaw jerks and orofacial myoclonia and another posterior variant with visual symptoms and alexia or dyslexia. We present a case with suggestible evidence of both conditions coexisting within the same patient, a finding that, to our knowledge, has not been previously reported. The diagnosis in this specific case was contributed to by the patient searching the internet.

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1. Introduction

Most episodes with loss of consciousness (LOC) are usually due to a banal syncope. The physician should always attempt to exclude potentially fatal conditions like e.g., long QT syndrome. We describe a male patient in his twenties with a rather unusual cause of LOC that was not especially conspicuous at presentation.

2. Medical history

The patient had repeated febrile seizures as a child. He was involved in a car accident in 2009 with following neck pain and consulted a physiotherapist. In 2011, he noticed intention tremor which also occurred in a brother and cousin. He was diagnosed with essential tremor. In the same year, he had an unspecified virus infection with fever, macular

erythema in one lower limb, itching, pharyngeal pain, headache, and signs of slight meningeal irritation.

3. First presentation and first round of diagnostic work-up

In 2012, he experienced his first episode with LOC while sitting on his balcony at home with a good book and coffee. He describes the onset where “the letters were dancing before his eyes with a consecutive blackout”. Loss of consciousness lasted for 5 min, and he was confused upon awakening. He noted slight pain in his forehead and his nose and knees were bruised. The mother observed jerking and stiffening in the extremities. In the ambulance, he experienced headache, nausea, and vomiting. There was no muscle pain. The neurologic examination, CT and MRI scan, ECG, EEG, and blood tests including CK were all normal. In the days preceding the LOC, he had been working many nightshifts. A few hours before the LOC, he ran a distance of 3000 m. The weather was rather warm for being in the autumn, with temperatures up to 28 °C and he did not properly rehydrate. The LOC was considered to be caused by sleep deprivation and dehydration. His previous medical history was not interpreted as relevant to the present event. The diagnosis was set as LOC possibly due to dehydration and, in any case, evaluated as a provoked seizure. His driving license was restricted temporarily.

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4. Visit in a private health institution and visit at the general practitioner

In 2013, he visited a private neurologist because of “jaw clicking” and involuntary movements in his jaw that preceded several occasions of LOC. Afterwards, he always felt tired and confused and malaise for several days. He started lamotrigine, but thereafter in his own words, he “turned into an analphabet and shuffled letters while reading”; further, he reported “dysphasia and stammering”. He was referred to the Department of Neurology. He then turned so photophobic that he mostly sat in a dark room with sunglasses on. He could not read a single sentence without experiencing his jaw clicking. He was not working and in the planning stages of education. The referral to the Department of Neurology was hastened.

5. New visit at the Department of Neurology and extensive diagnostic work-up

A polyclinic consultation revealed prodromal aura symptoms in the form of a burning sensation as well as an unpleasant feeling in his forehead and retroorbital location bilaterally during reading. This occurred especially while reading small text size and was possibly more pronounced while reading foreign languages. If he read for 10–20 s, jaw jerks appeared before he experienced further seizure progression. He experienced some symptom deterioration during sun exposure. There was no family history of epilepsy. He had no problems with regard to executive activities but experienced that caffeine at least partly could trigger seizures. The dose of lamotrigine was increased more rapidly, and he received add-on therapy with valproic acid. Extensive diagnostic

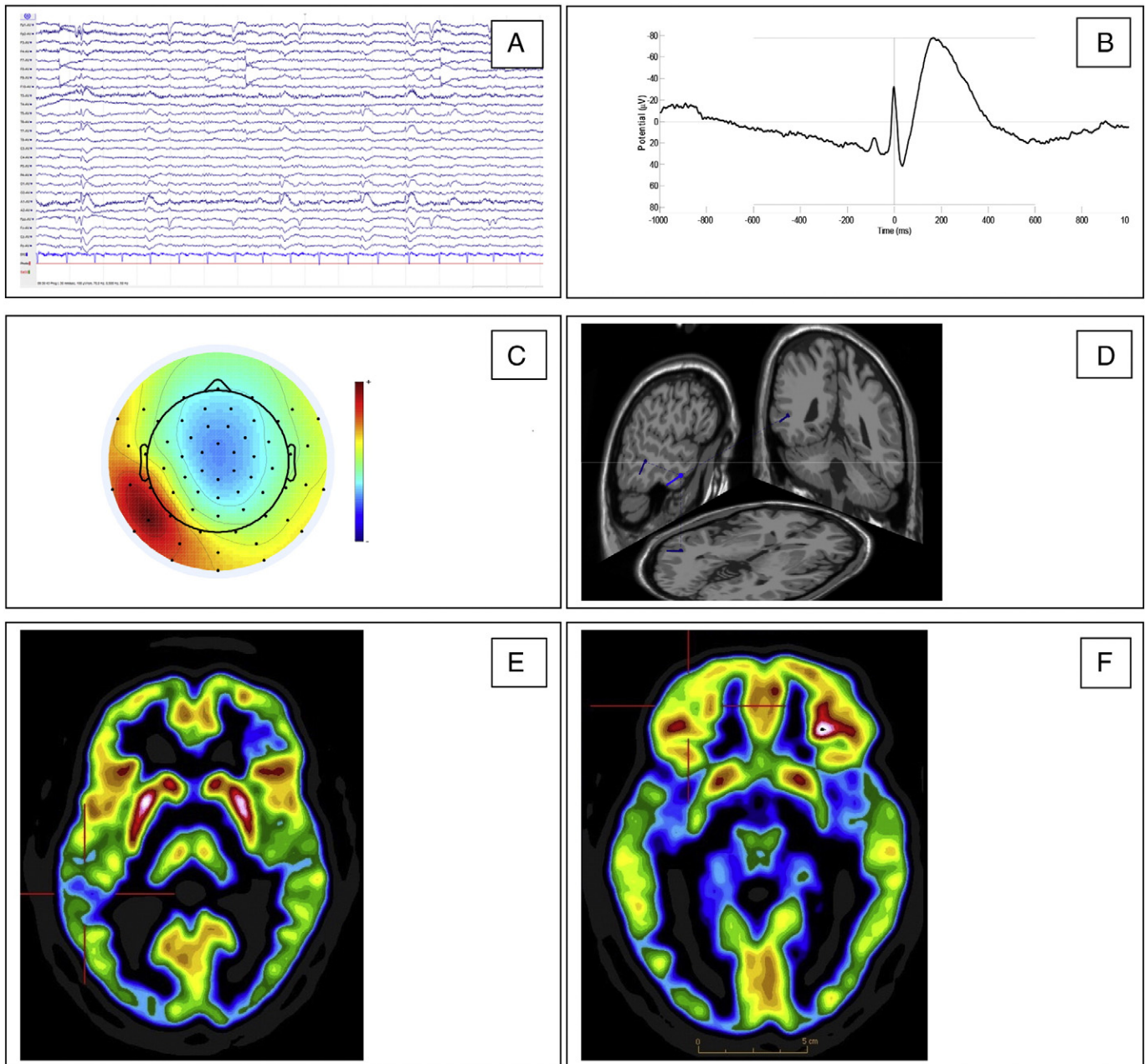


Fig. 1. A. Standard EEG reading English text. B. Serial spike slow wave averaging extracted from data by “template matching”. C. Topographic 2-dimensional map with spike localization from a 64-channel recording. D. Source analysis and localization of the spike generator. E. PET with asymmetric FDG-uptake in a temporooccipital localization in the left hemisphere. F. PET with asymmetric FDG-uptake in an orbitofrontal localization in the left hemisphere.

work-up was instigated; standard EEG recording with eyes open and eyes closed was normal, but when reading English (his second language) text, spike slow waves appeared in a left temporooccipital location (Fig. 1A). Three-Tesla MRI with epilepsy protocol was normal in 2012. This MRI was reevaluated in regard to an epileptic focus as found in EEG without any evidence of cortical dysplasia. The MRI 3D-reconstruction was normal in 2014, and this study was also interpreted negative for cortical dysplasia. In a 64-channel EEG while reading a foreign language text (English), a solitary jaw jerk appeared during the first of three reading sessions, and averaging of serial spike slow waves (Fig. 1B), topographic spike localization (Fig. 1C), and source analysis (Fig. 1D) were undertaken. A 32-channel EEG with simultaneous fMRI for 3D topographic mapping did not show any EEG–fMRI correlation. A PET scan showed asymmetric FDG-uptake in two different locations in the left hemisphere; temporooccipital (Fig. 1E) and orbitofrontal (Fig. 1F), which may indicate hypometabolism and dysfunction.

6. Discussion

The patient's first LOC fits reasonably well with heat cramps, but the seizures recurred. Several EEGs and cerebral images were all normal until the revelation appeared, or rightfully so, the patient realized the correlation with reading and insisted on reading during an EEG procedure, something he found out was necessary for the correct diagnosis while searching the internet for the words: "reading" and "seizures". Thus, the proper diagnosis is reflex-induced focal idiopathic partial epilepsy (International League Against Epilepsy, 1989).

Reading epilepsy usually presents itself in the 2nd or 3rd decade [1]. Reflex-induced seizures in reading epilepsy result when a critical mass of neurons is activated through a provoking stimulus within corticoreticular and corticocortical circuitry subserving normal functions [2]. The condition is part of a clinical spectrum with a most frequent variant clinically characterized by jaw jerks and a rare variant characterized by initial visual symptoms and a- or dyslexia (posterior variant) [1]. Both variants evolve to generalized seizures if the patient continues reading. A correct diagnosis depends on knowledge of the disease entity and sufficient anamnesis. Standard interictal EEG and cerebral imaging are usually normal. Ictal EEG shows bilateral synchronous spike slow waves in the variant with jaw jerks and temporoparietal focal spikes in the posterior variant [3]. Discharges tend to occur on the left (or dominant hemisphere) but can be bilateral or even switch sides [1,4,5]. Some patients have coexisting juvenile myoclonus epilepsy but then also exhibit spontaneous myoclonus in the morning [4].

Our case patient seems to possess components of both variants and can be interpreted to be an admixture in the abovementioned spectrum.

Interestingly, our patient did not have bilateral synchronous spike slow wave activity, but he had jaw jerks. The PET scan showed reduced metabolism not only in the left temporooccipital cortex but also in more frontal/orbitofrontal locations suggesting involvement of a larger network. The literature still points to a common pathophysiological substrate in both variants with involvement of posterior areas related to reading⁵, thus having one of the subtypes does not exclude the other.

Empirical treatment for the variant with jaw jerks is carbamazepine or valproic acid. Posterior variant is treated with antiepileptic medication as in simple partial seizures e.g., carbamazepine. As of the publication of this case report, he only takes valproic acid and has been seizure-free since 2013, but he has not been reading for almost two years.

Contributions

HKO – idea, manuscript preparation, interpretation, translation, literature search.

TE – idea, fMRI, literature search, multichannel EEG recordings, source analysis.

TS – PET scan and interpretation.

JL – MRI, fMRI interpretation.

IEH – idea, manuscript revision.

BAE – idea, manuscript revision.

Conflict of interest

None of the authors has any conflict of interest to disclose.

Written consent has been obtained from the patient.

We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this report is consistent with those guidelines.

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